Case Report

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An enormous benign primary retroperitoneal mucinous cystadenoma: a case report and literature review of a seldom seen abdominal pathology

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Introduction and importance: Retroperitoneal neoplasia comprise less than 2% of all tumours. benign primary retroperitoneal mucinous cystadenoma (PRMC) is an extremely rare tumour. Their clinical course is overall silent unless the patient presents with a vague abdominal or pelvic pain, abdominal distention, or a palpable mass. Their aetiology remains theorized and since 1989, only 46 cases (excluding ours) worldwide were documented in the literature. The majority of cases were discovered in females but the overall tumour incidence rate is still undetermined due to its rarity. Well-timed recognition of this pathology permits the necessary curative surgical intervention to take place.

Case presentation: We hereby illustrate the rare case of a 23-year-old female who presented to the surgical clinic complaining solely of an unexplained gradual increase of the abdominal contour. Their presurgical radiological analysis yielded an intraabdominal large-sized well-demarcated retroperitoneal mass.

Clinical discussion: Thorough resection of the mass was accomplished via open surgery. The subsequent microscopic analysis of excised tumour yielded the diagnosis of primary retroperitoneal mucinous cystadenoma of benign nature.

Conclusion: Primary retroperitoneal mucinous cystadenoma is a seldom seen tumour. The scarcity of its occurrence is further highlighted by the published data. Based on their conclusive review of the available published English-based literature, ours is the 47th documented case of a benign PRMC and it is the first documented case from our country; Syria. The impact of these findings warrants raising awareness on the subject and considering PRMC as a differential diagnosis when presented with a similar case in the clinical practice.

Keywords: abdominal mass, abdominal surgery, benign retroperitoneal tumour, case report, primary retroperitoneal mucinous cystadenoma

Introduction

Primary Retroperitoneal Mucinous Cystadenoma (PRMC) is a profoundly rare neoplasia. Despite showing gross and microscopic likenesses to the well-known cystadenomas that affect the ovaries, PRMCs originate and were found to be located in diverse retroperitoneal sites with zero anatomical relationship to the ovaries[1].

Based on the existing available literature, the number of documented cases PRMCs is especially low. Hence, the molecular and histopathological behaviour of these neoplasia remains highly reliant on theoretical hypotheses[2].

PRMCs resemble the majority of masses that are found in the retroperitoneum with regard to the symptoms reported by the affected patients. This similarity is expressed when these tumours

HIGHLIGHTS

- Retroperitoneal tumours are accountable for less than 2% of all types of neoplasia. The majority of which are malignant in nature.
- Primary retroperitoneal mucinous cystadenoma is an extremely rare tumour with poorly understood aetiological origins.
- The gold standard treatment approach remains utter surgical resection of the cystic mass.
- The definitive diagnosis can only be established after a thorough histopathological analysis of the resected specimen.
- Including this case, only 47 cases of a benign primary retroperitoneal mucinous cystadenoma have been documented in the English literature since 1989.

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begin to increase in size enough to instigate obstructions or pressure effects on the nearby structures and organs[3].

In terms of curative treatment for PRMCs, the current consensus recommends decisive and meticulous surgical resection of the tumour so that the possible subsequent ramifications, such as recurrence and infections, could be circumvented. Exploratory laparotomy allows for the optimal surgical field exposure and therefore, enables the utter removal and enucleation of the mass to take place while minimizing the risk of content spillage. Nonetheless, resection of PRMCs via laparoscopic approaches are documented in the literature[4,5].

After careful review of the available published English-based literature, we can conclude that ours is the 47th case of a benign PRMC worldwide since 1989 and the first one ever documented from our country. This magnifies the relevance and importance of our case to the scientific community.

The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines[6].

Presentation of case

Patient information

We present the case of a previously healthy 23-year-old Middle Eastern female who presented to our specialized General Surgery clinic complaining of a gradual but painless abdominal distention. The chief complaint began 2 months prior to her presentation. The abdomen was overall increasing in size and it was concurrently combined with a vague abdominal discomfort/feeling of heaviness.

It is valuable to mark that our patient denied experiencing nausea, vomitus, jaundice, appetite changes, bowel habits disruptions, dyspnoea, or genitourinary symptoms.

B-Symptoms such as fever, unintentional weight changes, or cold/night sweats were denied.

The patient's medical, surgical, gynaecological, drug, allergic, and psychosocial histories are all unremarkable. Furthermore, no previous similar complaints were reported.

The patient's BMI measured 21 Kg/m^2.

Clinical findings

The patient's vital signs were initially taken and recorded. No anomalous findings in their values were seen.

When inspecting the abdomen, it seemed to be generally distended. No skin changes, such as hypo/hyperpigmentation, spider naevi, caput medusa, pallor, or jaundice were seen. By abdominal palpation, the abdomen was soft and no tenderness or guarding was exhibited.

By percussion, the abdomen overall was slightly dull, whereas the auscultatory findings were unremarkable. The remainder of the physical examination did not yield any remarkable findings.

Diagnostic assessment

Our radiologic assessment was initiated by performing a transabdominal and pelvic ultrasound. An enormous-sized anechoic cystic formation with well-demarcated borders was seen extending throughout the abdomen and pelvis. It appeared to physically displace the bowel loops and the bladder.

To obtain better insight, the patient underwent a high-resolution multi-slice computed tomography (MSCT) scan of the chest, abdomen, and pelvis. An abdominal primary large-sized uninoculated cystic formation was found. It measured approximately ($22 \times 19 \times 13.5$ cm). It was situated in the left and middle sides of the abdomen and its borders were well-defined and the thickness of its wall reached up to (2.6 mm). It extended from the level of the left adnexa to the level of the inferior pole of the spleen (Fig. 1A-B-C). Furthermore, it seemed to push the small and large bowel loops and the left Fallopian Tube to the right side and the bladder inferiorly. The rest of the internal vital organs including the lungs (Fig.1 D) the liver, spleen, pancreas, kidneys, adrenal glands, inferior vena cava, and abdominal aorta were normal. Additionally, no lymphadenopathy or free fluid were seen. These findings propose the diagnosis of a retroperitoneal mucinous cystadenoma.

We drew blood samples for a complete laboratory analysis including testing for serum tumour markers like CA-125 and the results of which were all within normal limits.

Based on the previous givens, surgery was indicated. Thus, our patient was put on a nil-per-mouth nutritional status, we installed 2 large-bore intravenous cannulas to ensure adequate venous access, and she was administered the necessary preoperative antibiotics for prophylactic purposes as indicated by the healthcare guidelines.

No obstacles were come upon throughout any of the perioperative stages.

Therapeutic intervention

The gold standard treatment approach is the decisive and utter surgical resection of the lesion. We preferred relying on an open surgical pathway to avoid the risk of cystic rupture and spillage of contents into the abdomen especially because the cystic mass was considerably large in size. Exploratory laparotomy was carried out via midline incision at our specialized tertiary hospital under the umbrella of general anaesthesia. The operation was completed under the direct guidance and supervision of a General Surgery professor with 36 years of experience, by a General Surgery specialist and by a first surgical assistant with 10 and 5 years of experience, respectively.

No perioperative complications were reported.

Ideal surgical exposure of the field of the operation was obtained via a vertical midline abdominal incision. A giant cystic mass was immediately noticed. Its anatomical location was conformant with the findings of the previously mentioned MSCT scan. Meticulous isolation of the cystic mass from the surrounding peritoneal soft tissue was achieved to avoid spillage of its contents. It was then carefully removed from the abdominal cavity (Fig. 2A, B). No further findings were marked after a careful inspection of the abdominal vital organs and surrounding structures was done to ensure to additional lesions exist. The aforementioned cystic mass was immediately sent to our specialized histopathology laboratory to undergo a complete and meticulous analysis to establish a definitive diagnosis.

The outcome of which revealed the diagnosis of a benign primary retroperitoneal mucinous cystadenoma. Its contents were mucinous in composition, the internal and external surfaces were smooth with no vegetations, and the wall thickness ranges between (0.1 to 0.2 cm). No malignant features were demonstrated (Fig. 3A, B).

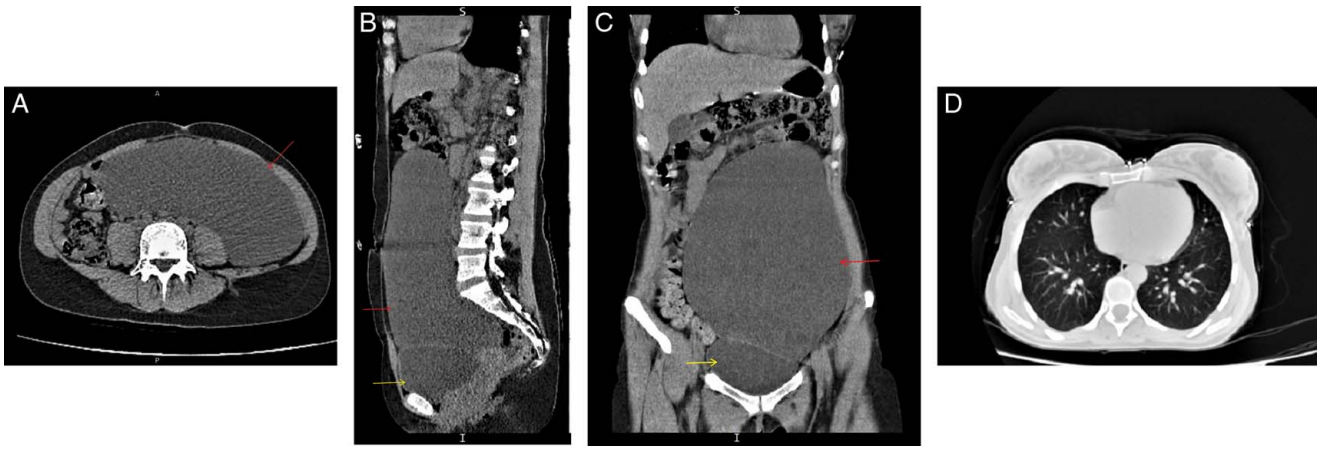

Figure 1. (A–C) Respectively, preoperative axial, sagittal, and coronal views of the Multi-Slice Computed Tomography (MSCT) scan of the chest, abdomen, and pelvis. The photos demonstrate a large-sized solitary well-demarcated uniloculated cystic formation that measures approximately (22 × 19 × 13.5 cm). The cystic content appears to be hypodense with an approximal density of 11 Hounsfield Unit. This cystic lesion occupies most of the abdominal space and is displacing the bowel loops in the left hypochondriac region toward the midline. It is also closely situated to the left psoas muscle and left iliac vessels. It is even compressing the nearby structures resulting in the displacement of the bladder inferiorly and anteriorly. The red arrow in the photos identifies the retroperitoneal cystic formation, whereas the Yellow Arrow identifies the bladder. (D): Preoperative axial view of the MSCT scan of the chest showing a clear lung field with no presence of metastasis from the abdominal cystic formation.

Our patient was discharged from the in-patient setting to her home within 2 days of the surgical intervention. Postoperative antibiotics were administered in addition to regular sterile wound dressings were applied to ensure the proper recovery of the wound. Moreover, she has been followed up by regular scheduled visits at our surgical clinic for 1 month thus far whereby she underwent conclusive laboratory, physical, and radiological assessments. All of which yielded normal results. Her path to a full recovery is so far lacking any obstacles.

Discussion

Based on the available scientific merit, it has been established that most retroperitoneal cystic masses are of malignant nature[7,8]. With that being said, PRMC is considered an epithelial tumour of benign nature[7]. Studies have shown that retroperitoneal neoplasia comprise less than 2% of all types of neoplastic occurrences. PRMCs are known to be extremely rare tumours and despite having a histological resemblance to ovarian mucinous cystadenomas, they are proven to possess the capability to originate from any site in the retroperitoneum without being anatomically related to the ovaries[9].

Hanfield-Jones was the pioneer who, in 1924, first depicted Primary Retroperitoneal Mucinous Cystadenomas in his paper that discussed cysts of the retroperitoneum[10]. In that study, he defined retroperitoneal cysts as follows: "*They are naturally situated behind the peritoneum, but I submit that the term should be reserved for those Cysts lying in the retroperitoneal fatty tissues which have no apparent connections with any adult anatomical structure*"

Regarding the histological origins of PRMCs, there are currently two chief hypotheses that are considered responsible. The first one argues that PRMCs are believed to be originating from heterotopic ovarian tissue due to its sound resemblance to ovarian cystadenomas. The second hypothesis considers PRMCs to originate from an invagination of the mesothelium of the peritoneum that becomes experiences mucinous metaplasia and forms as a cyst[11,12].

Relying on the available data on retroperitoneal tumours, they are sorted into three subtypes. The commonest of which is

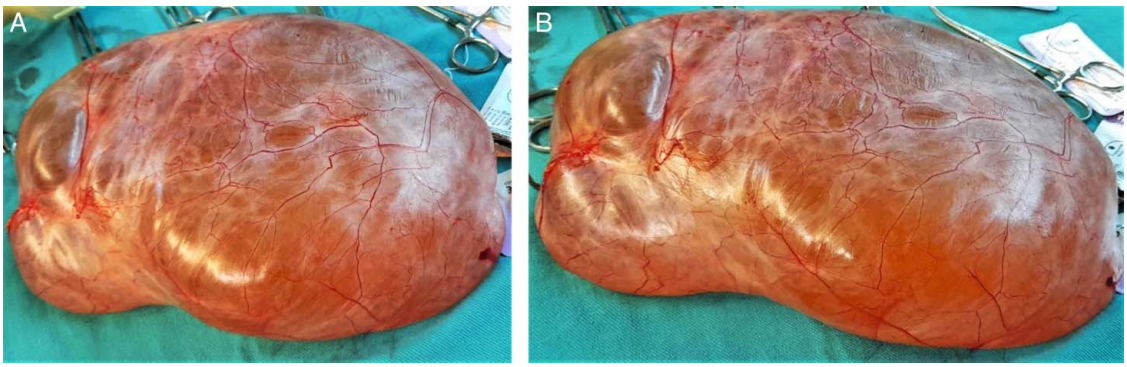

Figure 2. (A, B) Intraoperative images after the complete resection of the mass. It depicts the primary retroperitoneal mucinous cystadenoma.

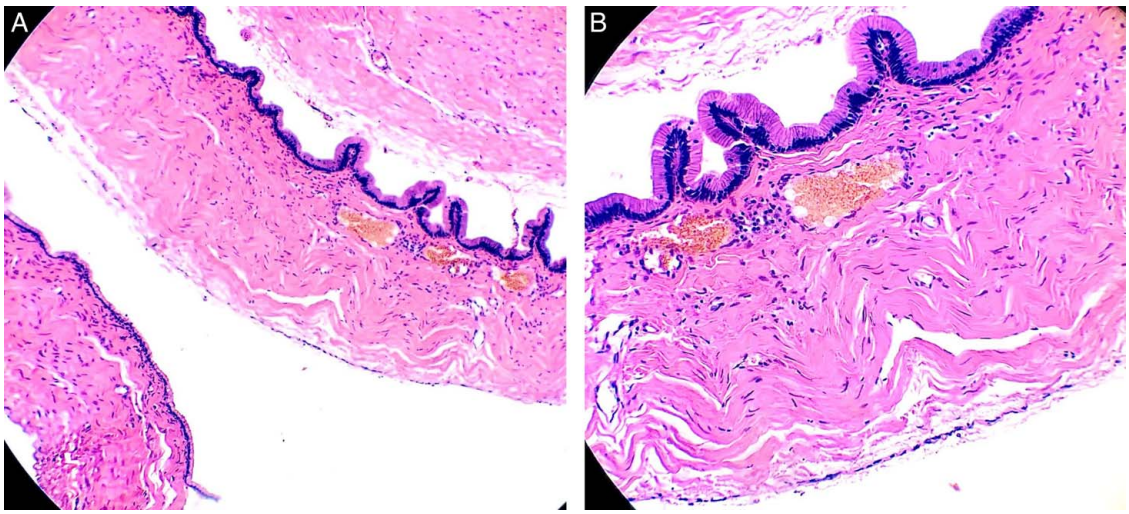

Figure 3. (A) Histopathological microscopic image of the resected cystic mass via Hematoxylin and Eosin (H&E) staining. The stroma consists of fibro-collagenous tissue with no ovarian components. (B) Histopathological microscopic image of the resected cystic mass via H&E staining. The cyst wall is lined with monolayered bland tall columnar cells with clear cytoplasm and basal nuclei (mucinous epithelium). No atypia or stromal invasion are noted.

retroperitoneal mucinous cystadenoma. This subtype is known to be benign and is not known to recur after complete excision. Furthermore, it appears as a large multilocular or unilocular cyst. The second subtype is similar to mucinous tumours of the ovary which have a low malignant transformation potential. Moreover, this type is characterized by the presence of foci of proliferating columnar epithelium within its lining. The third and final type is the malignant mucinous cystadenocarcinoma. In this type, segments of mucinous and benign tissue who possess a low malignant transformation capability in combination to a cystadenocarcinoma. Patients affected by this type are highly prone to experience postoperative tumour recurrence and their mortality rate is high due to metastasis[4].

Over the course of this tumour development, patients who are affected by PRMCs are archetypally asymptomatic. Moreover, these cysts are principally found as incidental findings during a routine medical evaluation. Since these tumours predominantly occur in females, such patients' most frequent chief complaint is non-specific or vague abdominal discomfort[13].

In terms of preoperative diagnostic approaches, a thorough transabdominal ultrasound examination is the first line of radiological investigations utilized to detect PRMCs as this is a readily available, economically efficient, and noninvasive diagnostic modality. Via ultrasound imaging, characteristics of malignant transformation could be investigated. These include increased wall thickness and septa formation. Nevertheless, ultrasound alone cannot optimally evaluate the lesion's origin or its local invasion of nearby organs[14].

We must bear in mind that detecting a retroperitoneal mass on ultrasonographical scanning warrants the performance of more complex diagnostic imaging to aid in further studying of the situation. Therefore, a CT scan is the contemporary gold standard diagnostic radiological tool that is done to aid in the diagnosis of these lesions. Furthermore, a CT scan can help in the evaluation of the following cystic characteristics: location, the presence of calcifications, the invasion of neighbouring structures, and necrosis. Additionally, it can study cystic wall details, such as wall thickness, integrity, regularity, and size[15]. The typical PRMC appearance demonstrated on abdominal CT scan is a unilocular homogenous cystic formation[16]. A vital point that provides us with clues that the above-mentioned mass is indeed retroperitoneal is the physical displacement of the abdominal organs, such as the ureters, colon, or kidneys[14,16].

To put the preoperative diagnostic tools into further perspective, analyzing blood samples for serum tumour markers and conducting analysis on the specimens of the PRMC's cystic fluid content are not found to be determinant factors toward establishing a definitive diagnosis of this neoplasia[14,16,17]. Further attempts to gain a clue on the diagnosis via aspiration of the cystic contents could yield in demonstrating the epithelial cell type and in turn, the histological class. Nevertheless, this invasive investigation is mostly avoided for two main rationales. First, because of its low specificity and sensitivity. Second, because it holds the devastating risk of cystic content spillage into the peritoneal space resulting in a life-threatening ramification known as Pseudomyxoma Peritonei or in case of malignant nature, tumour cell seeding[18,19].

With all that being said, the definitive diagnosis can only be accurately reached after meticulous histopathological analysis of the resected specimens[20].

The current gold standard therapeutic approach for PRMCs is total surgical excision of the mass so that the potential complications, such as infections and recurrence, could be safely evaded. Surgery in the form of exploratory laparotomy enables the surgeon to obtain optimal surgical exposure of the field and allows for the conclusive resection and enucleation of the tumour to be achieved. This helps avoiding content spillage into the abdominal space. Nevertheless, cases of PRMCs that were managed via a laparoscopic approach were documented in the available literature[4,5].

Upon careful review of the published literature, we report that Pespane and colleagues described 37 cases of PRMCs upon reviewing the literature from 1989 until May of 2017. During their search, they utilized the following keywords; primary, retroperitoneal, cystic adenoma, cystadenoma, and mucinous in the abstract, title, and/or keywords via Google Scholar, Medline, and

Scopus databases[21]. Furthermore, Foula *et al.*[15] reported an additional five cases—including theirs—of a benign PRMC documented between 2017 until 2019. In turn, we have found four more cases of benign PRMC between the years 2020 and 2023 utilizing the same above-mentioned criteria. These four cases are in chronological order as follows: Lung J. *et al.*[11], Danen C. *et al.*[22], Dorji N. *et al.*[23], Frini S. *et al.*[24]. As a result, we can conclude that ours is the 47th case of a benign PRMC worldwide since 1989 and the first one ever documented from our country.

Conclusion

Benign primary retroperitoneal mucinous cystadenoma is a tremendously rare neoplasia. The scarcity of its occurrence is additionally emphasized by the corresponding published data. Based on our meticulous review of the available published English-based literature, ours is the 47th documented case of PRMC worldwide and it is the first ever documented case from our country. The magnitude of these findings warrants raising awareness on this topic and demands considering PRMC as a possible differential diagnosis when presented with a comparable case in the clinical settings. Raising awareness to this pathology will contribute to considering it a differential diagnosis, thus decreasing the instances of misdiagnoses, save time for patients and physicians alike, and yield in performing timely therapeutic interventions that result in resolving this problem for our affected patients.

Future research prospects on this topic will aid in constructing clear and concise patterns and protocols for preoperative diagnosis, intraoperative management techniques and modalities, and postoperative patient follow-up guidelines. Finally, apt documentation of these cases is crucial to circumvent misdiagnosis. This helps plunge the rates of morbidity for the affected patients and helps establish proper preoperative clinical, surgical, and follow-up protocols for such patients.

Ethics approval and consent to participate

Institutional review board approval is not required for deidentified single case reports or histories based on institutional policies.

Consent of patient

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contributions

O.A.: Conceptualization, resources, methodology, data curation, investigation, who wrote, original drafted, edited, visualized, validated, literature reviewed the manuscript, and the corresponding author who submitted the paper for publication.

A.Ay.: Pathological analysis of resected specimens, assignment of the final histopathological diagnosis, validation, and review of the manuscript.

A.Ad.: Radiological analysis of the preoperative images, validation, and review of the manuscript.

R.S.: 1st surgical assistant during the operation, data curation, resources, validation, visualization, writing, and reviewing the manuscript.

A.Al.: General Surgery PhD student and specialist who performed and supervised the operation, in addition to supervision, project administration, and review of the manuscript.

H.H.: General Surgery professor who supervised the operation, in addition to supervision, project administration, and review of the manuscript.

All authors read and approved the final manuscript.

Conflicts of interest disclosure

N/A.

Research registration unique identifying number (UIN)

N/A.

Guarantor

Omar Al Laham.

Availability of data and materials

The datasets generated during and/or analyzed during the current study are not publicly available because the data were obtained from the hospital computer-based in-house system. Data are available from the corresponding author upon reasonable request.

Provenance and peer review

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